Case Reports

Coronary Artery Aneurysm - Two Case Reports

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Abstract:
Coronary aneurysms are defined as a localized dilatation that exceeds 1.5 times the diameter of the adjacent segment of artery. Aneurysms can be saccular (transverse larger than the longitudinal axis) or fusiform (longitudinal at least twice the transverse axis). These should be distinguished from coronary artery ectasia in which there is diffuse dilatation involving greater than 50% of the coronary artery.

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Introduction:
Coronary aneurysms are defined as a localized dilatation that exceeds 1.5 times the diameter of the adjacent segment of artery. Aneurysms can be saccular (transverse larger than the longitudinal axis) or fusiform (longitudinal at least twice the transverse axis).1 These should be distinguished from coronary artery ectasia in which there is diffuse dilatation involving greater than 50% of the coronary artery. Morgagni described the first case of coronary artery aneurysms in 1761 and Munker et al. reported the first antemortem case diagnosed by coronary angiography in 1958. Based on several angiographic studies, the incidence of coronary artery aneurysms ranges widely from 0.3% to 5.3% of the population, and a pooled analysis reports a mean incidence of 1.65%.2 A study from India reported an incidence of 10–12%, the highest in the literature to date, perhaps reflecting a specific genetic and/or environmental predisposition.3 The right coronary artery is the most commonly affected (40–87% of aneurysms), followed by the left circumflex or left anterior descending artery, depending on the study.4

Three-vessel or left main involvement is rare. Treatment options consist of surgical, percutaneous, and medical approaches. The largest experience in adults has been with surgical management, which typically includes bypass grafting. Percutaneous treatment is a newer option, with a markedly smaller data set, and includes stenting and coiling. In this case presentation, we will be showing two different treatment choices as per the angiographic characteristics.

Case study – 1:
A 45 yrs old diabetic, ex smoker male was admitted with acute inferior wall STEMI of 14 hrs duration. He neither had any H/O angina or SOB in past nor any other conventional coronary risk factors.

On admission, the patient was hemodynamically unstable, having a BP of 86/56 mmHg with features of peripheral hypoperfusion. His ECG showed ST elevation in II, III and aVF with ST depression in anterior precordial leads. Echocardiography revealed severe hypokinesia of inferior and inferoseptal walls with mild hypokinesia of anteroseptal wall, and the global LV function was around 40%. Considering the condition, he was immediately taken to cath lab for urgent revascularisation.

CAG revealed – ectasia in Proximal RCA with tight stenosis immediately distal to it causing TIMI I flow in distal part (Fig.-1 & 2) a large aneurysm in proximal LAD with 99% stenosis beyond it (Fig.-3). Ostial LAD and OM1 branch also showed significant lesions.

Management - considering the anatomy (multiple areas of aneurysm), the gentleman was taken up for urgent CABG with hemodynamic support.

Follow up – after complete revascularization, patient’s condition stabilized with few initial hiccups in post operative periods which were managed successfully.
Case study – 2:
A 55 year old nondiabetic, hypertensive female without any other conventional atherosclerotic risk factors, was admitted with acute inferior wall AMI and cardiogenic shock, almost 30 hours after onset of chest pain.

On admission, the patient had persistent hypotension needing high doses of inotropes. She was also having complete heart block for which a temporary pacemaker had to be inserted. Her chest examination showed evidence of pulmonary oedema. ECG was suggestive of IWMI with possibly RCA being the culprit. Echocardiography showed akinetic inferior wall, inferoseptal and anteroseptal wall with overall LVEF of 36%.

CAG revealed – ecstatic proximal LAD with midpart cut off. There was a giant aneurysm in proximal RCA of 30 X 22 mm size with distal part nonvisualised.

Management - since the RCA was thought to be unsuitable for percutaneous intervention, and the morbid state of the patient, it was decided to open up the LAD which was totally occluded from midpart, to tide over the crisis of shock. LAD flow was established percutaneously and the patient was sent for early CABG and aneurysmectomy.

Follow up - the patient performed better after complete revascularization. The heart block also disappeared.

Discussion:
Coronary artery aneurysm (CAA), defined as dilatation of the coronary artery exceeding 50% of the reference vessel diameter, is uncommon and occurs in <5% of coronary angiographic series. CAAs are termed giant if their diameter exceeds the reference vessel diameter by >4 times or if they are >8 mm in diameter. The difference between ectasia and aneurysm are often subtle and mainly semantic. If the length of the dilated segment is more than 50% of artery, it is called ectasia.

Congenital coronary artery aneurysms are rare. The most common etiology is atherosclerosis, present in 50–80% of cases. Histologic examinations of coronary aneurysms have revealed findings of arteriosclerosis, such as hyalinization and lipid deposition of the intima, intramural hemorrhage, and inflammatory reactions consistent with the arteriosclerotic process. The pathophysiology of aneurysm formation has been postulated to be weakening of the medial layer of the vessel wall, which in part may be due to chronic overstimulation of the vasodilator nitric oxide. Coronary aneurysms may occur from trauma (blunt and iatrogenic in the catheterization lab) and are the hallmark of Kawasaki disease. In the study by Swayne et al., patients with coronary aneurysms had similar coronary risk factors and no difference

Fig.-1: Prox RCA aneurysm with subtotal obstruction in LAO cranial view.

Fig.-2: Same findings in RAO cranial view.

Fig.-3: RAO caudal view showing large aneurysm in prox LAD and OM1
Coronary aneurysms are frequently seen in association with atherosclerosis, suggesting an overlap in risk factors and pathogenesis. It has been estimated that 50% of coronary aneurysms are due to atherosclerosis. Observations have led to the hypothesis that coronary aneurysms are a manifestation of atherosclerosis, with a similar pathophysiology. In support of this, patients with heterozygous familial hypercholesterolemia, who are at markedly increased risk for atherosclerosis, have a significantly increased incidence of coronary artery ectasia (15% vs. 2.5%).

The next most common cause is congenital, accounting for 20–30% of coronary aneurysms. A host of inflammatory and connective tissue disorders have also been associated with coronary aneurysms. Most well known is the association with Kawasaki disease, but coronary aneurysms have also been reported in patients with Takayasu’s arteritis, lupus, rheumatoid arthritis, Marfan syndrome, and Ehlers-Danlos syndrome. Coronary aneurysms have also been noted in conjunction with infection, drug use, trauma, and percutaneous coronary intervention.

The majority of coronary aneurysms are asymptomatic. When symptoms occur, angina or infarction are the most common presenting features. Myocardial infarction is the initial presentation in 30–50% of cases. Aneurysms may be complicated by thrombosis and rupture. Rupture is a rare event with no occurrences in the 978 patients with aneurysms in the CASS database. Thrombosis is more common. A postmortem study by Daoud et al. reported the presence of thrombus in 7 of 10 patients with coronary aneurysms. Several case reports have also demonstrated myocardial infarctions resulting from thrombosed coronary aneurysms without an obstructive coronary lesion. Thrombosis and distal embolization are thought to be potential etiologies of the clinical presentation. Swanton et al. have demonstrated reduced coronary blood flow in patients with coronary aneurysms, which may be a potential nidus for clot formation.

Demopoulos et al. reported ischemic event rates (cardiac death, myocardial infarction, or unstable angina) among 121 patients with concomitant coronary aneurysms and obstructive coronary disease (defined as the presence of >70% stenosis) versus 115 coronary disease patients with no aneurysms. There was no difference in event rates (12.4% vs. 10.4%; p = NS). Even though there is a recognized association between aneurysms and coronary atherosclerosis, the current case illustrates that a non-atherosclerotic coronary aneurysm can cause an ischemic event without an obstructing coronary lesion.

Treatment options consist of surgical, percutaneous, and medical approaches. Operative therapy may also include aneurysm ligation, resection, or marsupialization with interposition graft, and the ideal approach has not yet been formally studied. Percutaneous treatment is a newer option, with a markedly smaller data set, and includes stenting and coiling. The largest experience in adults has been with surgical management, which typically includes bypass grafting. Operative therapy may also include aneurysm ligation, resection, or marsupialization with interposition graft, and the ideal approach has not yet been formally studied.

Based on these limited data, the authors suggest that polytetrafluoroethylene-covered stents should be limited to patients whose aneurysms are <10 mm in diameter. Other important considerations associated with use of these stents include decreased flexibility, making implantation in tortuous vessels complex, and blocking access to side branches.

Percutaneous application of polytetrafluoroethylene (PTFE)-covered stents has gained popularity due their ability to effectively limit the expansion of CAAs by reducing blood flow within the aneurysm, thereby preventing their rupture. Some authors have suggested that PTFE-covered stents should be limited to patients whose aneurysms are <10 mm in diameter.

Percutaneous strategies also include coil embolization, autologous saphenous vein covered stent grafting, and one case report of DES implantation superimposed on a PTFE covered stent graft.

There are few data regarding medical therapy for coronary aneurysms. Medical management generally includes antiplatelet and/or antithrombotic agents, the use of which has been anecdotal. No data are currently available to indicate the relative merits of either approach.

The optimal imaging and management of giant CAA remain controversial and are based largely on case reports and anecdotal experience. Coronary angiography has some limitations, including the inability to differentiate true aneurysms from pseudoaneurysms or complex plaques. IVU provides detailed, high-quality images and is a valuable tool to distinguish CAA from coronary artery ectasia, as well as true aneurysms from pseudoaneurysms. Moreover, it is helpful in choosing appropriate stent dimensions for a therapeutic intervention and in ensuring full stent expansion after deployment. Cardiac computed tomography enables the
further delineation of the topographical anatomy of CAA and appears particularly useful during follow-up imaging after CAA exclusion with PTFE-covered stents, as illustrated in the present case.

Although our knowledge of coronary artery aneurysms and their management has progressed, a great deal remains unknown. Aneurysms are most commonly associated with ACAD, but it is not clear which factors promote the development of aneurysms in these patients. With regard to outcome, there is a discrepancy in the published series in how patients with aneurysms alone fare. It will be important to determine if these patients do better than those with aneurysms and concomitant ACAD, to formulate an appropriate treatment plan. For physicians managing patients with coronary artery aneurysms, it remains unclear whether anticoagulation and/or antiplatelet therapy offers benefits. Certainly, patients with concomitant ACAD should be treated with standard antiplatelet and lipid-lowering therapies to recommended prevention targets. Answers to questions regarding optimal treatment may come from newer clinical series or the development of a multicenter registry, as older published reports included many patients who did not have access to the full array of medical, percutaneous, and surgical options available today. With the increase in angiography and the more widespread use of high resolution CT scans and MRI imaging, the diagnosis of coronary aneurysms is likely to become more frequent, and the need for evidence-based management strategies will grow. As our knowledge of the pathophysiology and natural history of these lesions expand, treatment and outcomes for patients with coronary artery aneurysms are likely to continue to improve.

**Conflict of interest:** None

**References:**